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NATIONAL GUIDELINE CLEARINGHOUSE

General

Guideline Title

Congress of Neurological Surgeons systematic review and evidence-based guidelines on the role of imaging in the diagnosis and management of patients with vestibular schwannomas.

Bibliographic Source(s)

Dunn IF, Bi WL, Mukundan S, Delman BN, Parish J, Atkins T, Asher AL, Olson JJ. Congress of Neurological Surgeons systematic review and evidence-based guidelines on the role of imaging in the diagnosis and management of patients with vestibular schwannomas. Neurosurgery. 2018 Feb 1;82(2):E32-4. PubMed

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

NEATS Assessment

National Guideline Clearinghouse (NGC) has assessed this guideline's adherence to standards of trustworthiness, derived from the Institute of Medicine's report Clinical Practice Guidelines We Can Trust.

= Poor = Fair = Good = Very Good = Excellent

Assessment	Standard of Trustworthiness
YES	Disclosure of Guideline Funding Source
	Disclosure and Management of Financial Conflict of Interests
	Guideline Development Group Composition
YES	Multidisciplinary Group

UNKNOWN	Methodologist Involvement
▮▯▯▯▯	Patient and Public Perspectives
	Use of a Systematic Review of Evidence
▮▮▮▮▮	Search Strategy
▮▮▮▮▮	Study Selection
▮▮▮▮▯	Synthesis of Evidence
	Evidence Foundations for and Rating Strength of Recommendations
▮▮▮▯▯	Grading the Quality or Strength of Evidence
▮▮▮▮▯	Benefits and Harms of Recommendations
▮▮▮▮▮	Evidence Summary Supporting Recommendations
▮▮▮▯▯	Rating the Strength of Recommendations
▮▮▮▮▮	Specific and Unambiguous Articulation of Recommendations
▮▯▯▯▯	External Review
▮▮▮▯▯	Updating

Recommendations

Major Recommendations

Definitions for the classification of evidence (I-III) and levels of recommendations (1-3) are provided at the end of the "Major Recommendations" field.

Question 1

What sequences should be obtained on magnetic resonance imaging (MRI) to evaluate vestibular schwannomas before and after surgery?

Target Population

Adults with vestibular schwannomas.

Recommendations

Initial Preoperative Evaluation

Level 3: Imaging used to detect vestibular schwannomas should use high-resolution T2-weighted and contrast-enhanced T1-weighted MRI.

Level 3: Standard T1, T2, fluid attenuated inversion recovery, and diffusion weighted imaging MR sequences obtained in axial, coronal, and sagittal plane may be used for detection of vestibular schwannomas.

Preoperative Surveillance

Level 3: Preoperative surveillance for growth of a vestibular schwannoma should be followed with either contrast-enhanced 3-dimensional (3-D) T1 magnetization prepared rapid acquisition gradient echo (MPRAGE) or high-resolution T2 (including constructive interference in steady state [CISS] or fast imaging employing steady-state acquisition [FIESTA] sequences) MRI.

Postoperative Evaluation

Level 2: Postoperative evaluation should be performed with postcontrast 3-D T1 MPRAGE, with nodular enhancement considered suspicious for recurrence.

Question 2

Is there a role for advanced imaging for facial nerve detection preoperatively (e.g., CISS/FIESTA or diffusion tensor imaging)?

Target Population

Adults with proven or suspected vestibular schwannomas by imaging.

Recommendation

Level 3: T2-weighted MRI may be used to augment visualization of the facial nerve course as part of preoperative evaluation.

Question 3

What is the expected growth rate of vestibular schwannomas on MRI, and how often should they be imaged if a "watch and wait" philosophy is pursued?

Target Population

Adults with suspected vestibular schwannomas by imaging.

Recommendation

Level 3: MRIs should be obtained annually for 5 yr, with interval lengthening thereafter with tumor stability.

Question 4

Do cystic vestibular schwannomas behave differently than their solid counterparts?

Target Population

Adults with vestibular schwannomas with cystic components.

Recommendation

Level 3: Adults with cystic vestibular schwannomas should be counseled that their tumors may more often be associated with rapid growth, lower rates of complete resection, and facial nerve outcomes that may be inferior in the immediate postoperative period but similar to noncystic schwannomas over time.

Question 5

Should the extent of lateral internal auditory canal involvement be considered by treating physicians?

Target Population

Adult patients with vestibular schwannomas.

Recommendation

Level 3: The degree of lateral internal auditory canal involvement by tumor adversely affects facial nerve and hearing outcomes and should be emphasized when interpreting imaging for preoperative planning.

Question 6

How should patients with neurofibromatosis type 2 (NF2) and vestibular schwannoma be imaged and over what follow-up period?

Target Population

Adult patients with NF2 and vestibular schwannomas

Recommendation

Level 3: In general, vestibular schwannomas associated with NF2 should be imaged (similar to sporadic schwannomas) with the following caveats:

> More frequent imaging may be adopted in NF2 patients because of a more variable growth rate for vestibular schwannomas, and annual imaging may ensue once the growth rate is established.
> In NF2 patients with bilateral vestibular schwannomas, growth rate of a vestibular schwannoma may increase after resection of the contralateral tumor, and therefore, more frequent imaging may be indicated, based on the nonoperated tumor's historical rate of growth.
> Careful consideration should be given to whether contrast is necessary in follow-up studies or if high-resolution T2 (including CISS or FIESTA-type sequences) MRI may adequately characterize changes in lesion size instead.

Question 7

How long should vestibular schwannomas be imaged after surgery, including after gross-total, near-total, and subtotal resection?

Target Population

Adult patients with vestibular schwannomas followed after surgery.

Recommendation

Level 3: For patients receiving gross total resection, a postoperative MRI may be considered to document the surgical impression and may occur as late as 1 yr after surgery. For patients not receiving gross total resection, more frequent surveillance scans are suggested; annual MRI scans may be reasonable for 5 yr. Imaging follow-up should be adjusted accordingly for continued surveillance if any change in nodular enhancement is demonstrated.

Definitions

American Association of Neurological Surgeons/Congress of Neurological Surgeons Classification of Evidence on Diagnosis and Levels of Recommendation

Class I Evidence Level 1 Recommendation	Evidence provided by one or more well-designed clinical studies of a *diverse* population using a "gold standard" reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.
Class II Evidence Level 2 Recommendation	Evidence provided by one or more well-designed clinical studies of a *restricted* population using a "gold standard" reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.
Class III Evidence Level 3 Recommendation	Evidence provided by expert opinion or studies that do not meet the criteria for the delineation of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Vestibular schwannomas

Guideline Category

Diagnosis

Evaluation

Management

Clinical Specialty

Neurological Surgery

Neurology

Otolaryngology

Radiology

Intended Users

Physicians

Guideline Objective(s)

To critically analyze the primary literature regarding the role of imaging in the management of vestibular schwannomas (VSs)

Target Population

- Adults with proven or suspected vestibular schwannomas (VSs)
- Adults with neurofibromatosis type 2 (NF2) and VSs

Interventions and Practices Considered

1. Magnetic resonance imaging (MRI)
2. Counseling
3. Preoperative surveillance
4. Posttreatment surveillance intervals
5. Imaging caveats for vestibular schwannoma associated with neurofibromatosis type 2 (NF2)

Major Outcomes Considered

- Sensitivity and specificity of magnetic resonance imaging (MRI) techniques
- Tumor recurrence rates
- Tumor growth rates
- Facial nerve function
- Hearing preservation rates

Methodology

Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Hand-searches of Published Literature (Secondary Sources)

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Process Overview

The evidence-based clinical practice guideline taskforce members and the Joint Tumor Section of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) conducted a systematic review of the literature relevant to the management of vestibular schwannomas (VSs). Additional details of the systematic review are provided below and within the introduction and methodology chapter of the guideline (see the "Availability of Companion Documents" field).

Study Selection and Eligibility Criteria

A total of 2070 citations were manually reviewed. Two independent reviewers evaluated and abstracted full-text data for each article. Citations focused on the imaging of VSs in adult patients largely in the magnetic resonance imaging (MRI) era (January 1, 1990 to December 31, 2014), published in English, were considered.

Investigated patients suspected of having VSs
Patients ≥18 years of age
Was of humans
Published between January 1, 1946 and December 31, 2014
Quantitatively presented results
Was not an in vitro study (for novel molecular markers, in vitro studies were included on patient samples)
Was not a biomechanical study
Was not performed on cadavers
Was published in English
Was not a meeting abstract, editorial, letter, or commentary
Studies may include mixed pathology, however the data pertaining to acoustic neuromas (ANs)/VSs was abstractable from the paper
>5 patients or patient samples

Systematic reviews, guidelines, or meta-analyses conducted by other authors were not included in this guideline creation. These documents were developed using different inclusion criteria than those specified in this guideline. Therefore, they may have included studies that do not meet the inclusion criteria stated above.

Search Strategies

The task force collaborated with a medical librarian to search for articles published between January 1, 1990 and December 31, 2014. The following electronic databases were searched: PubMed and Cochrane Central. Strategies for searching electronic databases were constructed by the evidence-based clinical practice guideline taskforce members and the medical librarian using previously published search strategies to identify relevant studies (see Figure 1 and Table 1 in the full guideline [see the "Availability of Companion Documents" field]). The guideline committee also examined lists of included and excluded studies for errors and omissions.

Number of Source Documents

Ninety-six studies were included as evidence. See Figure 1 in the full guideline (see the "Availability of Companion Documents" field).

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

American Association of Neurological Surgeons/Congress of Neurological Surgeons Classification of Evidence on Diagnosis and Levels of Recommendation

Class I Evidence Level 1 Recommendation	Evidence provided by one or more well-designed clinical studies of a *diverse* population using a "gold standard" reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.
Class II Evidence Level 2 Recommendation	Evidence provided by one or more well-designed clinical studies of a *restricted* population using a "gold standard" reference test in a blinded evaluation appropriate for the diagnostic applications and enabling the assessment of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.
Class III Evidence Level 3 Recommendation	Evidence provided by expert opinion or studies that do not meet the criteria for the delineation of sensitivity, specificity, positive and negative predictive values, and, where applicable, likelihood ratios.

Methods Used to Analyze the Evidence

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

Data Collection Process and Assessment of Bias

Abstracts that met the inclusion criteria were retrieved in full text form and evaluated for confirmation that they met criteria as suggested by prior abstract review. The information was then used for construction of the evidence tables.

The possibility of systematic bias in results was addressed by first stratifying the evidence based on the class of evidence quality, which highlights the limitations in this literature. Given the dearth of evidence for many of these questions, formal methods for studying publication bias, such as funnel plots were not

feasible.

In addition, one obvious bias inherent to these studies is selection bias. For a patient to be in an imaging study, that patient, by definition, underwent imaging for a clinical reason, which may bias results toward larger and possibly more aggressive tumors than would be seen in a cohort of all vestibular schwannomas (VSs). However, it is important to note that this bias is uniform across all studies of this type. Therefore, while individual practitioners may have skewed results by differences in case selection, there is no clear mechanism by which these biases are systematically distributed.

Methods Used to Formulate the Recommendations

Expert Consensus (Nominal Group Technique)

Description of Methods Used to Formulate the Recommendations

Classification System and Recommendation Formulation

The concept of linking evidence to recommendations has been further formalized by the American Medical Association (AMA) and many specialty societies, including the American Association of Neurological Surgeons (AANS), Congress of Neurological Surgeons (CNS), and the American Academy of Neurology (AAN). This formalization involves the designation of specific relationships between the strength of evidence and the strength of recommendations to avoid ambiguity. Refer to the "Rating Scheme for the Strength of the Evidence" field.

Guideline Panel Consensus

Multidisciplinary writing groups were created for each section based on author expertise to address each of the disciplines and particular areas of therapy selected for these clinical guidelines. Each group was involved with literature selection, creation and editing of the evidence tables, and scientific foundations for their specific section and discipline. Using this information, the writing groups then drafted the recommendations in answer to the questions formulated at the beginning of the process, culminating in the clinical practice guideline for their respective discipline. The draft guidelines were then circulated to the entire clinical guideline panel to allow for multidisciplinary feedback, discussion, and ultimately approval.

Rating Scheme for the Strength of the Recommendations

See the "Rating Scheme for the Strength of the Evidence" field.

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Internal Peer Review

Description of Method of Guideline Validation

The completed evidence-based clinical practice guidelines for the management of vestibular schwannomas (VSs) were presented to the Joint Guideline Committee (JGC) of the American Association of Neurological Surgeons (AANS) and the Congress of Neurological Surgeons (CNS) for review. The reviewers for the JGC

were vetted by *Neurosurgery* for suitability and expertise to serve as reviewers for the purposes of publication in that journal also. The final product was then approved and endorsed by the executive committees of both the AANS and CNS before publication in *Neurosurgery*.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate evaluation and surveillance of vestibular schwannomas

Potential Harms

At the time of publication, the evolving concerns around gadolinium retention within the brain and nephrogenic systemic sclerosis raise the consideration of avoiding contrast altogether if the overarching goal of routine surveillance is to identify lesion growth.

Qualifying Statements

Qualifying Statements

Disclaimer of Liability

This clinical systematic review and evidence-based guideline was developed by a multidisciplinary physician volunteer task force and serves as an educational tool designed to provide an accurate review of the subject matter covered. These guidelines are disseminated with the understanding that the recommendations by the authors and consultants who have collaborated in their development are not meant to replace the individualized care and treatment advice from a patient's physician(s). If medical advice or assistance is required, the services of a competent physician should be sought. The proposals contained in these guidelines may not be suitable for use in all circumstances. The choice to implement any particular recommendation contained in these guidelines must be made by a managing physician in light of the situation in each particular patient and on the basis of existing resources.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Quick Reference Guides/Physician Guides

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

Dunn IF, Bi WL, Mukundan S, Delman BN, Parish J, Atkins T, Asher AL, Olson JJ. Congress of Neurological Surgeons systematic review and evidence-based guidelines on the role of imaging in the diagnosis and management of patients with vestibular schwannomas. Neurosurgery. 2018 Feb 1;82(2):E32-4. PubMed

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2018 Feb 1

Guideline Developer(s)

Congress of Neurological Surgeons - Professional Association

Source(s) of Funding

These evidence-based clinical practice guidelines were funded exclusively by the Congress of Neurological Surgeons and the Tumor Section of the Congress of Neurological Surgeons and the American Association of Neurological Surgeons, which received no funding from outside commercial sources to support the development of this document.

Guideline Committee

Vestibular Schwannoma Evidence-Based Practice Guideline Task Force

Composition of Group That Authored the Guideline

Task Force Members: Ian F. Dunn, MD, Department of Neurosurgery, Brigham and Women's Hospital, Harvard Medical School, Boston, Massachusetts; Wenya Linda Bi, MD, PhD, Department of Neurosurgery, Brigham and Women's Hospital, Harvard Medical School, Boston, Massachusetts; Srinivasan Mukundan, MD, PhD, Division of Neuroradiology, Brigham and Women's Hospital, Boston, Massachusetts; Bradley N. Delman, MD, Department of Radiology (Neuroradiology), Icahn School of Medicine at Mount Sinai, New York, New York; John Parish, MD, Carolinas Medical Center, Charlotte, North Carolina; Tyler Atkins, MD, Carolina Neurosurgery and Spine Associates, Charlotte, North Carolina; Anthony L. Asher, MD, Carolinas Medical Center, Charlotte, North Carolina; Jeffrey J. Olson, MD, Department of Neurosurgery, Emory University School of Medicine, Atlanta, Georgia

Financial Disclosures/Conflicts of Interest

Conflict of Interest

The Vestibular Schwannoma Guidelines Task Force members were required to report all possible conflicts of interest (COIs) prior to beginning work on the guideline, using the COI disclosure form of the American Association of Neurological Surgeons/Congress of Neurological Surgeons (AANS/CNS) Joint Guidelines Committee, including potential COIs that are unrelated to the topic of the guideline. The CNS Guidelines Committee and Guideline Task Force Chair reviewed the disclosures and either approved or disapproved the nomination. The CNS Guidelines Committee and Guideline Task Force Chair are given latitude to approve nominations of Task Force members with possible conflicts and address this by restricting the writing and reviewing privileges of that person to topics unrelated to the possible COIs. The conflict of interest findings are provided in detail in the full-text introduction and methods manuscript (see the "Availability of Companion Documents" field).

Guideline Endorser(s)

American Association of Neurological Surgeons - Medical Specialty Society

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Guideline Availability

Available from the Neurosurgery Web site .

Availability of Companion Documents

The following are available:

Congress of Neurological Surgeons systematic review and evidence-based guidelines on the role of imaging in the diagnosis and management of patients with vestibular schwannomas. Full guideline. Schaumburg (IL): Congress of Neurological Surgeons (CNS); 2017 Dec 22. 78 p. Available from the

Congress of Neurological Surgeons (CNS) Web site .
Congress of Neurological Surgeons systematic review and evidence-based guidelines on the treatment of adults with vestibular schwannomas: introduction and methods. Schaumburg (IL): Congress of Neurological Surgeons (CNS); 2017 Dec 22. 28 p. Available from the CNS Web site .
Olson JJ, Kalkanis SN, Ryken TC. Congress of Neurological Surgeons systematic review and evidence-based guidelines on the treatment of adults with vestibular schwannomas: executive summary. Neurosurgery. 2018 Feb 1;82(2):129-34. Available from the Neurosurgery Web site .
Congress of Neurological Surgeons (CNS). Guideline development methodology: endorsed by the American Association of Neurological Surgeons (AANS), the Congress of Neurological Surgeons (CNS), and the AANS/CNS Joint Guideline Committee. Schaumburg (IL): Congress of Neurological Surgeons (CNS); 2012 Feb. 12 p. Available from the CNS Web site .

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on May 7, 2018. The information was verified by the guideline developer on June 4, 2018.

This NEATS assessment was completed by ECRI Institute on April 25, 2018. The information was verified by the guideline developer on June 4, 2018.

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